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REVIEW ARTICLE

Long-term outcome of male genital reconstruction in childhood☆

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Abstract Hypospadias, epispadias with or without exstrophy, and disorders of sex development are among the most common anomalies of genitalia that occur during childhood. Considering the tremendous effect of genitourinary reconstruction on adult life, the evaluation of the long-term results of different techniques of genitoplasty in pediatrics is of the utmost importance.

After reviewing the literature, the authors summarize the available long-term outcomes of genitoplasty in childhood, specifically focusing on the cosmetic, psychosocial, psychosexual and functional results, and emphasize that, contrary to the widely available data on early outcomes of genital reconstruction in the pediatric population, very few well described controlled studies have evaluated the long-term effect of genitoplasty in puberty and adulthood, in the sense that the surgeon should describe the peroperative findings in more detail and also be more structured in evaluating the postoperative result at follow-up visits.

Finally, the authors conclude that more attention should be paid to the impact of these techniques on cosmetic aspects and psychosexual development in these patients after puberty, as they play a crucial role in their adult quality of life.

Introduction

External genital reconstruction in pediatrics is challenging. Hypospadias, epispadias with or without exstrophy, and disorders of sex development are among the most difficult problems encountered by pediatric urologists. They require a lot of experience and are best treated by a multidisciplinary approach [1]. In this group of patients, management

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is complex and there is much disparity concerning the surgical correction. More than 300 different techniques with a wide variety of modifications have been introduced for the treatment of these anomalies [1,2]. Concerning the tremendous effect of genitourinary reconstruction on adult life, the evaluation of the long-term results of different techniques of genitoplasty in pediatrics is of the utmost importance. The complications of these reconstructive techniques can take decades before becoming evident [3,4]. Pubertal growth can alter the final functional and cosmetic aspects of the corrected genitalia. Moreover, psychosexual development is only completed after puberty, so the psychological and sexual function of patients who have undergone genital reconstruction can only be evaluated after puberty [1,5—7]. Considering the fact that as the patients grow up the cosmetic aspect becomes equally if not more important than function, more attention should be paid to the esthetic results in the long term [8,9]. Genital and reproductive functions have a great effect on the quality of life of adult patients with penile anomalies in childhood [7]. However, most published studies have reported only short-term results in pre-pubertal patients, with very few reports of long-term results focusing on the cosmetic and psychosexual outcomes of these surgeries in adulthood.

The objective of this review is to present the available long-term outcomes of genitoplasty in childhood, specifically focusing on cosmesis, and the psychosocial and functional results. The results should help pediatric urologists to decide how the current surgical techniques for genitoplasty meet the long-term goals for genitourinary reconstruction, and to inform the parents of newborns with penile anomalies about future expectations.

Hypospadias

Hypospadias is one of the most frequent problems in pediatric urology, occurring in approximately 1 out of 200 to 1 out of 300 live male births; however more recent studies suggest that the incidence of this anomaly is increasing [10—12].

Up to now several classifications have been introduced for hypospadias [13]. These classifications have considered the location of the meatus (before and after chordee correction), the glans (cleft, incomplete cleft or flat), the quality and width of the urethra, the prepuce (incomplete or complete), the degree of penile curvature and the presence of scrotal transposition [14,15]. Although many categorizations have been proposed for hypospadias to make it easy for surgeons to decide on the severity of this anomaly, it's believed that the severity of hypospadias can only be judged during surgery when the penis is degloved and the corpus spongiosum is clearly observable. We believe that the severity should be established during surgery, and that observation through dissection is the most important factor in making the best choice of procedure. As the division of corpus spongiosum is usually more proximal than the urethral opening, the position of the meatus is a poor criterion for judging the severity of hypospadias [16].

The only treatment for hypospadias is the surgical correction of this anomaly. Over the past century, many techniques and procedures have been introduced, which confirms the fact that there is no gold standard technique for this anatomical defect [17,18]. Most of these surgical procedures follow the same steps including correction of a ventral curvature, urethral reconstruction, and penile skin reconstruction. The current goals in the treatment of hypospadias are: (1) creating a straight penis, with (2) a terminally situated slit-like meatus (that allows for a full urinary stream and normal coitus), (3) normalization of erections and voiding, (4) creating a urethra of satisfactory and uniform caliber, and (5) symmetrical appearance of the glans [19—21].

During the past decades the major trends in the management of hypospadias have changed. During the first half of the 20th century, multi-stage repairs were considered as gold standard treatment. In the 1980s, with the advent of microsurgical techniques, new techniques of handling tissues with great care, and new absorbable suture material, one-stage repair was advised by many urologists for hypospadias repair. Recently this trend has been challenged by newly developed techniques that involve mobilization and transection of the urethral plate and ventral lengthening of the corpus cavernosa, which have popularized two-stage approaches again [22,23]. Determining the best technique depends on several factors including: surgeon experience, meatal position, the meatus and glans configuration, presence or absence of chordee, quality of ventral skin coverage, quality of the urethral plate, the availability of foreskin and the circumcision status. Snodgrass and colleagues ascertain that the proper technique can be only assessed when the lateral and ventral portions of the penis have been completely dissected and three factors are identifiable: 1) the level of corpus spongiosum diversion, 2) urethral plate quality, and 3) the length of urethra that needs reconstruction [16]. The quality of the urethral plate is usually determined by the length, width and thickness of the urethral plate during surgery. The thickness of the urethral plate is an important factor for the success of tubularized incised plate procedures, whereas the width of the urethral plate plays a significant role if a longitudinal incision is needed. Snodgrass believes that if one surgeon could preserve the urethral plate during the orthoplasty a tubularized incised plate urethroplasty is indicated. One may have a normal glans and meatus, but have an abnormality along the penile shaft with a poorly developed corpus spongiosum that is called chordee without hypospadias. Glans configuration and size is another important factor that every surgeon should take into consideration before choosing the technique. Patients with shallow or absent glanular groove require incisions or flap procedures [16].

Although the impact of hypospadias continues through adulthood and new techniques are continuously evolving, only a few reports have discussed the long-term outcomes of hypospadias repair. Due to the rapid growth at puberty, there is a potential risk of new problems; for example, a new asymptomatic micro fistula may start leaking, or the neourethra might fail to grow adequately during puberty causing new curvature, or the penile shape and length may cause concerns [1,5—7]. In this review, we outline the available long-term data under three categories: micturition, cosmesis and psychosexual. Table 1 presents the available long-term studies on hypospadias repair.

Table 1 Included hypospadias repair papers and summary of results.

First author	Year	Technique	Mean age at operation (years)	Mean age at follow up (years)	Total number
Bracka [19]	1989	Ventralizing, Terminalizing	5	22	196
Aho [24]	1997	Mathieu, Denis-Browne, Ombredanne	3.8	29.6	43
Aho [25]	2000	Mathieu, Denis-Browne, Ombredanne	3.6	29.5	48
Hoag [26]	2008	Meatal tube, Byars flap, MAGPI	1.95	20	28
Moriya [41]	2006	Mathieu, Duplay	3.8	20.6	22
Moriya [27]	2007	Mathieu, Duplay	3.8	20.6	22
Rynja [28]	2009	Mathieu	2.3	22	66
Goyal [32]	2010	Oral mucosa graft	7.5	18	30
Orkiszewski [37]	2004	Longitudinally neo-urethra	N/A	15—45	9
Patel [34]	2004	Onlay island, Island tube	1.4	16.6	30
Mureau [39]	1996	van der Meulen repair	N/A	N/A	35
Vandendriessche [40]	2010	N/A	N/A	15.6	10
Jiao [43]	2011	Bladder mucosa graft, Duckett technique, Thiersch-Duplay technique, Mathieu technique	6.37	21.6	43
Aulagne [44]	2010	Duckett tube, Ceci-culp, Cahuzac	3.3	25	27
Lam [45]	2005	Two staged	1.7	15.6	27
Ververidis [46]	2005	Snodgrass technique, Mathieu repair	N/A	N/A	32
Jones [29]	2009	Single stage, two stage	1.96	N/A	55
Seibold [49]	2010	MEMO technique	4.9	11.5	99
Bubanj [51]	2004	N/A	N/A	27.8	37

Micturation

In one of the first studies on the long-term outcome of hypospadias repair, Bracka revealed that patients who have undergone hypospadias repair in childhood experience more complications in the long term. He observed that 38% of his patients were dissatisfied, with spraying during voiding [19]. Aho et al. were among the first to report long-term micturition outcomes in their series of 43 and 48 patients who had undergone the Mathieu technique in childhood. The mean patient age at last follow up was 29 years. They observed that 80.5% of patients were experiencing lower urinary tract symptoms and 78% were dissatisfied with their urinary function. Spraying (63%), post-void dribbling (29.2%) and urinary stream deviation (26%) were among the most common micturition abnormalities reported in these studies [24,25]. They observed that controls were more satisfied with their urinary function. This observation was confirmed by the recent long-term studies by Hoag, Moriya, and Rynja et al. [26—28]. New studies have demonstrated that patients with severe hypospadias will experience more episodes of spraying compared to patients with distal hypospadias [29]. In the study by Rynja et al. it is observed that patients with proximal hypospadias experience fewer micturition problems compared to patients with distal hypospadias in the long term. There is a lot of disagreement among different studies in this area which reveals the need for more controlled studies with long-term follow up [28]. Manzoni et al. suggest that patients who underwent buccal mucosa graft repair should have regular follow up after puberty, as there is little known about the long-term behavior and growth of the buccal mucosa, which is less androgen sensitive than genital tissue [30]. Neslon et al. were among the pioneers who published their data on long-term outcomes in patients with oral mucosa graft urethroplasty for hypospadias. They observed a 20% risk of lower urinary tract symptoms and 4% risk of severe urinary symptoms; 26% of patients in this group complained of urinary spraying in the long term [31]. In a recent study, Goyal et al. noted very low complications with oral mucosa graft for hypospadias repair after puberty. They observed that only 10% of patients complained of lower urinary tract symptoms including urinary stream deviation, spraying and prolonged voiding [32].

There are a few studies which have used uroflowmetry for evaluation of micturition problems after hypospadias correction in the short term and long term. Most studies with long-term follow up with uroflow patterns revealed a maximum flow rate (Q_{max}) ranging from 8.4 to 31.1 ml/s [28,33—35]; however Rynja et al. in their recent study noted that only patients with proximal hypospadias have lower mean Q_{max} compared to control patients [28]. Most of these studies observed that in about 3.7%—14.7% of patients the Q_{max} values were lower than the 95th percentile of the normograms [28,29,33,36]. However, Orkizewski et al. observed that 44.4% of their patients who had undergone a TIP procedure have a Q_{max} lower than the 95th percentile [37]. The severity of hypospadias (penoscrotal hypospadias in all patients) and absence of spongious tissue could explain these findings. In 2004, Patel and colleagues reported the long-term follow up of 30 patients out of a cohort of 125 patients; 14 patients had undergone tubularized transverse island flap repair and 16 had undergone an onlay procedure. All patients in this follow up had normal urine flows with all Q_{max} values higher than the 95th percentile [34]. The 6.5 years' follow up of patients with

single-stage genital skin graft urethroplasty advocate that uroflow parameters remain stable during long-term follow up ($Q_{max} = 30.8 \pm 10.2$, $Q_{max} = 29.9 \pm 8.9$; 3 month and 6 years respectively; $P = 0.64$) [38]. Finally, from the few available controlled studies, what can be concluded is that patients with a previous history of hypospadias surgeries are less satisfied with their micturition in comparison to controls, and that this necessitates further investigations and even further evolutions in hypospadias surgery.

Cosmesis

Nowadays, there is a general agreement that hypospadias correction should result in a normal appearing penis with a slit-like meatus on the glans penis. There are very few studies that have assessed cosmetic results beyond the subjective impressions of surgeons [10]. The objective determinants of cosmesis in a patient with hypospadias can include a standardized scoring system with or without photographs that can be used by healthcare providers. Although objective assessment of cosmetic appearance is not that easy as patients do not have much to compare. In a long-term report of 196 patients (mean age at follow up 22 years) who had undergone hypospadias repair, Bracka noted that 30.6% of patients were dissatisfied with penile appearance and penile curvature was observed in 19.8% of patients [19]. In contrast, a study by Murau et al. revealed that 81% of patients were dissatisfied with their penile appearance after puberty, which could be attributed to an operation technique that is no longer used [39]. In 2005, Nelson and colleagues published their study of 65 patients who had undergone single-stage buccal mucosa urethroplasty. With a mean follow up of 7 years they observed that 28% of their patients were very satisfied and 12% were dissatisfied with penile appearance [31]. Two studies have been recently published which have assessed the long-term cosmetic outcomes in patients who had undergone hypospadias surgery in childhood compared with age-matched normal subjects [40,41]. In the first study, the authors observed that 40% of adolescents with hypospadias and 35% of controls were dissatisfied with penile appearance but there was no significant difference ($p = 0.184$). The only reason for dissatisfaction with penile appearance in this group of patients was smaller penile size, whereas curvature, phimosis and smaller penile size were among the main reasons in the control group [41]. In the other study, fewer boys with hypospadias surgery in childhood considered their penis to be normal or judged their penis as similar to that of friends in comparison with the normal control group. This group used the Junior Genital Perception Scale to evaluate the perception of body image in both groups. They noted that hypospadias patients were less satisfied with their body image compared to the control group (16.4 vs. 12.4 $p = 0.02$) [40]. Several studies have proven that patients with severe hypospadias are less satisfied than hypospadias patients in general [28,41,42]. Jiao et al. noted that patients with proximal hypospadias were more vulnerable to accept further cosmetic surgeries than those with distal hypospadias in adulthood [43].

The recent study by Alugne et al. [44] revealed that a slit-like meatus was achieved in 55% of patients with proximal hypospadias who had undergone the Duckett tube technique; whereas Lam et al. [45] noted that a slit-like meatus was observable in all patients with a history of two-stage repair in childhood.

In a study by Ververidis et al. the cosmetic result of the Snodgrass technique was compared with the meatal-based flap and onlay island flap repair in 32 boys who had their penis photographed following these procedures. The photographs were evaluated by a panel of five healthcare professionals. The mean evaluation score for meatus, glans, shaft and overall appearance was higher for the Snodgrass technique ($P < 0.05$) [46].

The Pediatric Penile Perception Score (PPPS) and the Hypospadias Objective Scoring Evaluation (HOSE) are the only two published evaluation instruments that have been used for assessment of anatomical long-term outcome [28,29,44]. The PPPS has 4 main domains: meatus, glans, shaft skin and general appearance, with a high score of 12 [47]. Rynja et al. assessed patient anatomical outcomes in the long term using the PPPS and they noted a PPPS of 8.5; however there is not a clear cut-off point for normal anatomical outcome [28]. The HOSE instrument includes 5 domains, comprising meatal location and shape, urinary stream, curvature at erection, and fistula. A HOSE score of more than 14 is defined as normal anatomical outcome [48]. In a study by Jones et al., HOSE was used to evaluate the long-term outcome in boys treated for hypospadias in childhood: 80% of patients had an excellent surgical outcome during adolescence [29]. These findings are confirmed by a recently published study of long-term outcomes of a mobilization technique: 94% of patients reached the maximum of 16 points while 97% achieved an excellent surgical outcome [49].

Psychosexual

There has been much controversy concerning the psychosocial and sexual outcomes following hypospadias correction. Svensson et al. are among the pioneers who investigated the late sexual effects of previous hypospadias surgery in adulthood [50]. They interviewed 34 men operated upon in childhood for hypospadias and 36 men operated upon for appendicitis at the same time for past and present social, sexual and psychological concerns. No significance difference was noted between the two groups regarding age at first ejaculation but hypospadias patients reported a later first intercourse in comparison with the control group. They also observed that hypospadias patients had a lower capacity for social and emotional relations than the controls. Interestingly, they did not find any significant correlation between severity of hypospadias and number of sexual partners, self-assessed sexual drive and number of coitions. Schonbucher et al. suggested that sexual satisfaction is generally lower among hypospadias patients compared to controls [51]. This finding was confirmed by Bubanj and colleagues, who observed that libido was the same in these patients compared to a control group of adult men, but that sexual function was significantly less satisfactory. They concluded that this could be attributed to low self-esteem and dissatisfaction with cosmesis [52]. These findings led to concerns about the

effect of hypospadias correction on ejaculatory and sensual functions of the penis. Recently, Rynja et al. noticed that frequency of ejaculation and orgasmic sensation after sexual stimulation was lower in hypospadias patients than controls. They measured erectile dysfunction by means of the International Index of Erectile Function in both groups. The mean score of the erectile domain was 20.6 in patients and 22.4 in controls, which did not reach the significance level. This finding was more prominent in patients with proximal hypospadias in childhood. They observed a lower frequency of masturbation in the hypospadias group than in controls, which is not confirmed by other studies [28]. In another study, which evaluated sexual function after oral mucosa graft urethroplasty, the authors noted that all 10 patients were satisfied with their erectile function with a erectile domain score of 26.8, and all experienced an orgasmic sensation most or all of the time, although 1 never ejaculated and 2 ejaculated only rarely [31].

A new controlled study by Moriya et al. revealed that hypospadias patients reported erectile problems more often than the controls (73% vs. 45%, $p = 0.04$). They observed that sexual behaviors were similar in the two groups, which was consistent with previous studies [41]. Long-term studies appear to support that intercourse frequencies do not differ between patients with hypospadias and the normal population [25,44].

In the recent study reviewing 43 adult patients following severe hypospadias surgery, 17 had problems during ejaculation. In the overall evaluation of sexual function, 19 (44.2%) patients described their sexual function as completely satisfactory, and 7 (16.3%) were dissatisfied. Of the 18 patients who had experienced sexual intercourse, 88.9% reported no problem, while 11.1% had difficulties during intercourse due to smaller penis size and severe curvature. In another study of 19 patients, 79% were satisfied with their erection and orgasm; however only 36% reported satisfactory ejaculation. The main complaints were weak ejaculation or the need to milk the urethra [43].

Most studies have confirmed that the level of intimate relationships is not different among hypospadias patients compared to controls [28,41,52]. However, some studies have proposed that patients with severe hypospadias may have a higher rate of avoiding sexual relationships. In a study by Bubanj and colleagues it was noted that the average total number of coital partners was statistically lower in the hypospadias group compared to controls, and this was more prominent in patients with distal hypospadias. These findings could be generally related to negative genital appraisal [52]. According to these available data, one may conclude that controls have greater satisfaction with their sexual function in adulthood than patients who had undergone hypospadias surgery. However, data on the effect of hypospadias and its reconstruction on the sexual behavior and sexual function of adult patients with hypospadias are very scarce, and further controlled studies with direct interviews are needed to evaluate the psychosexual aspects of hypospadias repair in childhood. One may argue that hypospadias is an isolated anomaly, and long-term follow-up patients may be limited to those who had complaints about this anomaly in adulthood. What is obvious from these studies is that the reporting of hypospadias patients dissatisfied with their sexual and micturitional function and their cosmetic appraisal of the genitalia necessitates prospective studies with long-term follow up of all patients who had hypospadias surgeries in childhood to draw a better conclusion on the future of hypospadias surgery.

Exstrophy/epispadias complex

The exstrophy—epispadias complex (EEC) is a rare congenital anomaly with an incidence of 1:10,000 to 1:50,000 and a 2:1 male:female ratio [53]. This anomaly represents a spectrum of abdominal and pelvic midline malformations ranging in severity from epispadias (presenting as the mildest form) to classic bladder exstrophy and cloacal exstrophy (as the most severe form) [54—56].

The treatment of children with EEC remains one of the most challenging areas in the field of reconstructive urology. Historically, reconstructive surgeries in this group of patients were focused on preservation of renal function and achievement of continence. Significant improvement in understanding of the embryology, anatomy and pathophysiology of this anomaly has led to noteworthy changes in the management of EEC during the past decades [2,57—59]. Recent advances in urinary continence in children with EEC has been achieved based on two main surgical techniques, the Jeffs and Gearhart and recently Grady and Mitchell, for one-stage complete repair of classic bladder exstrophy [59,60].

Associated genital anomalies in male patients with EEC include a short, flat, broad and tethered upward penis with the urethral opening along the dorsum of the penis and absent dorsal foreskin [8]. Previously, genitalia reconstruction was delayed until complete bladder closure and then until puberty; however, today it is usually performed together with the initial surgery in the neonatal period [1].

Regardless of the time when the genitoplasty is performed in this group of patients, it can have a dramatic effect on the patient's life in the long term. As these patients reach puberty, the cosmetic and esthetic aspects may play a greater role in well-being than does urinary continence [61]. However, there is a paucity of knowledge about the long-term outcome of genitoplasty in ECC patients with regard to genital function, psychosexual development and voiding function. We review the long-term outcomes of genital reconstruction in EEC patients from the published literature with regard to psychosexual aspects and cosmesis in adulthood. The available long-term studies of male genital reconstruction in EEC patients are outlined in Table 2.

Psychosexual development

Genital reconstruction in children with EEC can have tremendous long-term impacts on their self-esteem, sexuality, body image and relationships. Some studies have noticed that patients with ECC in childhood who had undergone genital reconstruction did not experience any psychosexual and psychosocial disturbance during their childhood [62]. However, Reiner and colleagues noted that all of their patients who underwent genitalia reconstruction reported a universal and chronic dysfunction and anxiety

Table 2 Included exstrophy/epispadias repair papers and summary of results.

First author	Year	Technique	Mean age at operation (years)	Mean age at follow up (years)	Total number
Ben-Chaim [62]	1996	N/A	N/A	N/A	16
Reiner [63]	1999	N/A	N/A	N/A	14
Diseth [64]	1998	N/A	N/A	14.5	22
Ebert [65]	2005	Single stage repair	N/A	N/A	54
Baird [66]	2004	N/A	3.1	35	7
Stein [67]	1994	N/A	N/A	N/A	33
Ebert [68]	2010	Single stage repair	2.6	23.4	17
Surer [69]	2000	Modified Cantwell-Ransley	3.4	N/A	12
Kibar [70]	2008	Complete penile disassembly	N/A	N/A	21

that had led to social and sexual developmental impairment [63]. The authors noted that only 2 out of 14 patients had ever undressed in front of others, only 2 had masturbated and only after 16, and only 2 had experienced sexual intercourse. One half of the patients had a mood disorder [63]. In another study by Diseth et al., 59% of patients were dissatisfied with their penile appearance. Sexual relationships were the main predictor of psychosocial dysfunction in their 15—20 year age group of patients with bladder exstrophy and epispadias. The authors concluded that the presence of a psychosocial expert in healthcare teams is necessary at all stages of EEC patients care [64]. The group from Germany evaluated the long-term psychosocial and psychosexual development of adolescents with EEC after complete repair. All patients rated their genital satisfaction and genital sensation as low and 58% were anxious about their sexual activity, whereas 43.9% had experienced sexual intercourse. They observed that anxiety about genital appearance was present even for patients with normal genitalia [65]. In a similar study, Baird et al. published results on 6 men with genital reconstruction for bladder exstrophy treated from 1960 to 1982. They found that 4 (66%) of them were capable of penetrative intercourse and ejaculation. They finally advocated that most patients with EEC repair in childhood are capable of acceptable sexual function when grown up [66].

Stein et al. looked at the long-term results of 33 male patients who had undergone bladder exstrophy or epispadias surgery with regard to social relationships, sexuality and fertility. Of these children, 28 had undergone genital reconstruction while 5 did not require genitoplasty. Eleven of the patients with genitalia reconstruction had a penile deviation that was troublesome in erection for two. None of the patients in the external genitalia reconstruction group could ejaculate normally or had achieved fatherhood, while all patients with no genital reconstruction in childhood had normal ejaculation and two had fathered children. The authors finally concluded that male patients with genital reconstruction have a potential risk of infertility and all parents should be informed about the risk of future infertility [67].

In comparison with the previous study, single-stage repair is a newer technique with few reports available yet on long-term outcomes. Ebert and colleagues presented 21 patients with EEC and a mean follow up of 19 years; 17 (81%) of these patients had undergone single-stage repair. They reported satisfactory erection in all patients and normal ejaculation in 19. Most patients (76%) had attempted sexual intercourse. They observed a high incidence of penile deviation in their patients that was attributed to the Young procedure; however, few patients complained about penile deviation during sexual intercourse and penile shortness was the most common annoying problem. Consequently, they advocated that single-stage repair will result in better sexual function in patients with EEC [68].

As Reiner has proposed, patients with EEC experience several issues during their life that can affect their developmental process [69]. These hurdles include: 1) imperfect urinary control in childhood, 2) complex mechanisms of bladder control, 3) unappealing appearance of genitalia, 4) malfunction of genitalia or malposition of genitalia, 5) higher prevalence of infertility. The author believes that these patients have a high prevalence of internalizing disorders over the period of their development. As there are no screening tools for psychosexual development in these children, the author highlights the role of the urologist in educating the parents and the child during the adolescent period to deal with their urinary insentience and genital abnormalities as the main constituents of psychosexual development. He believes that when sexual anxiety is intensified in these patients during adolescence, it results in avoidance of relationships and can even lead to severe disturbance of the developmental process that necessitates psychological assessment during this critical period of development. It can be said that patients with a history of genital reconstruction for EEC may experience some degree of sexual function impairment, although single-stage repairs could be more beneficial in these patients, but this should be confirmed by more long-term studies.

Cosmesis

Despite several long-term studies on the outcome of ECC repair in childhood regarding continence, quality of life and intercurrent diseases, very few studies have evaluated the outcomes of genital reconstruction in boys with ECC. Adolescents are more concerned about the appearance of their external genitalia than about continence [6]. Therefore, early efforts at esthetic reconstruction of genitalia may

relieve some their concerns in adulthood. In 2000, Surer et al. reported their 10-year follow up of 93 patients with a single-stage modified Cantwell-Ransley repair. In 93% of patients the penis was angled downward or horizontal while the patient was standing. Of the 12 patients who were older than 17 years old, 10 (83%) reported an acceptable cosmetic appearance [70]. In a recent study by VanderBrink et al. the long-term outcomes of primary and secondary genitalia reconstruction in 65 boys with EEC were reviewed [8]. Of these patients, 43 were born with classic bladder exstrophy and 22 with epispadias. In their series, 92% were satisfied with the esthetic outcome while 8% were dissatisfied and underwent esthetic revisions. They noted that the esthetic satisfaction rate was higher (94%) in patients who had undergone Cantwell-Ransley epispadias repair, despite more need of surgical revision due to the higher incidence of urethrocutanous fistula and urethral strictures. Hypospadias was the most frequent complication in patients who had undergone the Mitchell technique. They suggested using rotational flaps to improve cosmetic appearance [8]. In 2008, long-term results of the Mitchell technique for correction of epispadias showed that 33% of patients who had undergone complete penile disassembly were left with hypospadias, and that all of these cases were seen in patients with classic bladder exstrophy and none in patients with isolated epispadias [71].

Regarding the present data, there are considerable pitfalls in the long-term cosmetic follow up of patients with EEC. There is no questionnaire or valid instrument to report the esthetic outcome. There is no single study with more than 10 years' follow up that has evaluated the cosmetic outcomes in adulthood. The only conclusion is that the penile disassembly technique most often results in hypospadias and that long-term follow up and new approaches are needed to improve the cosmetic outcomes.

Possibly the revival of the Kelly technique would result in better esthetic outcomes; however there are no long-term data, to date. This technique is usually proposed as part of a staged closure in which a penoscrotal urethrostomy during penile lengthening is created and penoscrotal hypospadias is repaired at the age of 3. In contrast to the disassembly technique the meatus is left at the penoscrotal junction and not brought to the glans until a secondary reconstructive repair.

Disorders of sexual development (DSD)

Management of patients with DSD is challenging and genital reconstruction is essential for success [72]. In the past it was believed that gender reassignment was the best option in patients with severely undermasculinized genitalia due to the more successful surgical and endocrine feminization procedure. During the past decades significant changes have been made in surgical techniques for masculinizing genitoplasty. Recently, more concern has been raised regarding the in utero brain masculinization that can lead to gender dysphoria in adulthood. Schober et al. reported dissatisfaction in patients with a small phallus and perineoscrotal hypospadias who had undergone gender conversion [73]. So, nowadays, even children with small phallic structures and unresponsive to hormonal stimulation are not recommended for gender conversion [74]. Male genitoplasty in DSD patients who are assigned as males usually includes: chordee correction and penile straightening, urethroplasty, glanuloplasty, correction of scrotal deformities, and orchiopexy [75]. Despite the recent advances in surgical techniques for male genitoplasty in patients with DSD, there are several questions that are not well answered regarding these approaches. Will the reconstructed genitalia function normally and be well-adjusted when the patients grow up? What information can we provide about the functional and psychological outcomes in adulthood? There are very discrepant studies that have investigated the long-term outcomes of masculinizing surgeries in intersex patients (Table 3). Migeon et al. reported the long-term medical, surgical and psychosexual outcomes of 46XY individuals with ambiguous genitalia and perineoscrotal hypospadias [76]. Of the 39 patients, 21 were reared male and had undergone masculinizing genitoplasty. The appearance of external genitalia was reported using the Quigley grading scheme with a 7 point scale (1 = male phenotype to 7 = female phenotype). They noted a mean Quigley grade of 3.4 in this group of patients. The authors noted that 56% of patients had a penile length at or below 2.5 standard deviations of the mean norm established by Schonfeld and Beebe (mean stretched penile length = 8.8 cm). Six patients (28%) were completely satisfied with their sexual function while 14% were mainly dissatisfied. All patients in this series were capable of normal erection, and 90% reported the experience of sexual intercourse. Eleven patients were married or in long-term relationships and two had achieved fatherhood. Finally, the authors noted that 78% of the patients were satisfied with their gender of rearing. Sircili et al. looked at the long-term results of the Denis-Browne technique in their series of 59 patients with DSD and genital ambiguity who were raised as males. Morphological surgical results were acceptable in 97% of patients [77]. The mean penile length in 38 adult patients was 7.5 $\pm$ 2.1 cm at the last follow up. In contrast to the Migeon series they noted that only 2 patients had a penile length less than -2 SD.

Table 3 Included masculinizing genitoplasty in patients with DSD papers and summary of results.

First author	Year	Technique	Mean age at operation (years)	Mean age at follow up (years)	Total number
Migeon [74]	2002	N/A	N/A	N/A	21
Sircili [75]	2010	Denis-Browne technique	6	22	59

Patients with 5α-reductase deficiency had the smallest mean penile length while patients with testosterone deficiency showed the greatest mean penile length. Patients with partial androgen insensitivity experienced the least post-treatment penile length gain. The authors reported that 95% of patients were able to void standing and 73% had a good urinary stream without spraying. Of the 38 adults, 33(87%) were sexually active of whom only 9% were unsatisfied with their sexual activity. The authors did not find any significant relationship between the final penile length and patient satisfaction with sexual activity. Most patients in this series (89%) were satisfied with the anatomical and functional surgical results. The major complaints of patients who were dissatisfied with the surgical outcome were short penile length and difficult voiding due to urethral stenosis.

Regarding the present evidence, there is a scarcity of studies on the long-term effect of male genital reconstruction in patients with DSD, including patient appraisal of his genitalia or functional and cosmetic outcomes, which could possibly have tremendous effects on the current trends in management of DSD patients who undergo male gender reassignment. One may discuss that most available studies on long-term outcomes in DSD patients compare the outcomes between patients with different molecular backgrounds, while very few studies have evaluated the long-term outcomes of specific backgrounds of DSD in detail. Nonetheless, as genitalia reconstruction is a small component of a more generalized problem in this subgroup of patients, genitalia cannot be described as the only driving force in the overall dissatisfaction.

With the possibilities of phalloplasty and the excellent long-term cosmetic and functional outcomes in transsexuals, male gender assignment has to become the gold standard for undervirilized DSD patients [78,79].

Conclusion

There is a paucity of data on the long-term effect of different genital reconstruction techniques during childhood due to a lack of validated measurement tools and validated questionnaires, and high loss of patients during follow up. Very few studies have investigated the different outcomes of these surgeries in the long term especially after puberty and adulthood.

The studies available indicate that patients who underwent hypospadias surgeries are less satisfied with their urinary function in comparison with controls, and they usually experience spraying, post-void dribbling and urinary stream deviation, symptoms that are more prominent in patients with a history of severe and proximal hypospadias. Dissatisfaction about the sexual function and appearance of the penis is more prevalent in hypospadias patients than in controls, in adulthood. Although genital reconstruction in exstrophy patients impairs their sexual function in the future and some patients run the potential risk of infertility, a single-stage repair may result in better sexual function and patient satisfaction with their sexual activity. Very few studies have reported the long-term outcomes of genital reconstruction in DSD patients, but what can be noted is the high rate of satisfaction with their sexual function in adulthood and that the most frequent complaint among these patients is short penile length in comparison with the norm.

Regarding the presented data in this review, further controlled long-term studies are needed to select the best techniques for these groups of patients, as well as better consultation of parents with regard to the future of their children before advising on surgery. More attention should be paid to the impact of these techniques on psychosexual development and cosmetic aspect after puberty, as these factors play a crucial role in later quality of life in this subgroup of patients. Most of the studies reviewed in this paper do not report the mean number of follow-up years after the reconstructive surgery, and instead report the mean age at follow up as an indication of the postpubertal and long-term evaluation. This can be noted as a major critique, and authors of future studies need to pay attention to this defect. One point should be emphasized: that surgeons should describe the pre-operative findings in more detail and also be more structured while evaluating the post-operative results at follow-up visits. Without long-term follow-up evaluation of genital reconstructions in childhood with precise description of pre- and post-operative data, no technique can be considered as the gold standard in the management of patients with penile anomalies during infancy.

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